THE SURGICAL TREATMENT OF DYSTONIA MUSCULORUM DEFORMANS*

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The severe disability of patients afflicted with dystonia musculorum deformans arouses the urge to give them relief even at the cost of other neurologic sacrifices. Unfortunately, the etiology of the disorder is too poorly understood at present to permit a specific attack upon brain areas responsible for the abnormal motion.

The bizarre, yet stereotyped movements do not characterize a disease, but constitute a syndrome for which no specific pathologic lesion is known. The syndrome is considered to operate primarily via the extrapyramidal system. The abnormal involuntary motor activity is regarded as a release phenomenon, the result of loss of the inhibitory action of higher controlling centers. The motions persist too long to be reasonably attributed to an irritative lesion.

Dystonia consists of slow, long-sustained turning movements of the head and trunk, with rotation of especially the proximal parts of the extremities. In addition to these torsion movements, there are slow, sustained tensions in the platysma, shoulder muscles, pectorals and leg and foot muscles. Particularly long-sustained, simultaneous tensions of opposing muscle groups characterize dystonic movements. Physiologically, they resemble those of athetosis, but are longer sustained.

This report concerns the results obtained by surgical interruptions of pathways at several levels in the central nervous system for relief of severe dystonia in 3 children.

CASE REPORTS

Case 1. D.Y. was first seen at the age of 7 years. For a year, she had suffered from spasticity of both arms and from cramping spasms of the feet causing pain that made her cry at night. Within 6 months dystonic movements became pronounced.

Over the next 5 years the dystonia progressed. The head retracted, and the extremities were maintained in semiflexion and were relatively fixed. The facial muscles were not involved, but those of the trunk and extremities forced the body into grotesque postures. These movements were extremely painful and the child was reduced to an emaciated state. Extensive drug therapy was ineffective and she was admitted to the Children's Hospital at the age of 12 years to permit attempts at surgical relief, especially of her pain.

Operations. Encouraged by the results obtained in cases of unilateral Parkinsonism, on July 11, 1946 we first partly severed the anterior limb of the right internal capsule by the method of Meyers\textsuperscript{2} and Browder.\textsuperscript{1} This did not alter the patient's condition.

On July 22, 1946, the right area 6 was removed, sparing area 4. This abolished the dystonia and permitted weak voluntary motion of the hand. The right-sided dystonia increased, especially in the foot.

Fearing the effect of a bilateral cortical resection, since the child was intelligent, we sought control of the right side in an attack at the cord level. On July 30, 1946 the right anterior cord fasciculus was cauterized (Putnam's method\textsuperscript{4}) at C1–2 level. This left the child comfortable. The arms and legs were maintained flexed. Right grip was possible and the arm could be extended without eliciting dystonia. Involuntary movement was present in the right leg. Passive extension of the left arm resulted in a strong, coarse tremor and in flexion of the extremities. The neck still retracted to the left moderately under tension or emotion. A more complete denervation of the left side seemed desirable.

On Aug. 28, 1946 the right area 4 was excised above the level of lip response to stimulation. This resulted in a left hemiplegia, without tremor or reflex spasm.

In the next 9 months all gains were preserved; the child gained 20 pounds, was pain-free, friendly and comfortable in a wheelchair existence. Involuntary movement followed use of the right leg, which was flexed at knee and thigh. In an attempt to relieve this, the right pyramidal tract was severed\textsuperscript{5} at the 3rd dorsal level on May 12, 1947.

Course. Two years after the last operation (May, 1949) the 15-year-old girl was happy and pain-free. She was 50 pounds heavier than at the start of her surgical "career." Her I.Q. still measured 110 and she was a freshman in the Crippled Children's School. She subsequently has been graduated. Her head was tilted slightly to the right. The spine was in marked scoliosis. She could stand in braces, but could not walk. No dystonia nor resting tremor was present. Right grip was satisfactory and she could write legibly but slowly.

Within the last 3 years, coarse tremor and some dystonia have appeared in the right arm and have progressed until the extremity is almost useless. A left pedunculotomy is being considered to improve this.

Comment. This child has been observed over a period of 10 years. Primarily to relieve pain, successive surgical sacrifices were made to include finally: The right cortical areas 4 and 6, the right extrapyramidal tracts (anterior cord fasciculus) at C1–2 and the right pyramidal tract at D3 level. These resulted in abolition of the dystonia at the cost of a left hemiplegia and a right hemiparesis. The end result was happily accepted by the patient and her relatives.

Case 2. J.N. was a 9.5-year-old boy, admitted to the Children's Hospital on April 25, 1949. The patient was one of twins, the sister being normal. He weighed 3 pounds (1.4 kg.) at birth and lived in an incubator for 3 months. During infancy and after, he had difficulty in eating, eye movements were uncoordinated and his extremities displayed involuntary movements. Despite 4 years of intensive cerebral palsy management his condition deteriorated. For 8 months he had suffered frequent painful attacks of opisthotonos and flexion of the knees and hips.
Examination. He was poorly developed and nourished and weighed 34.5 pounds (15.7 kg.). He lay in bed in severe opisthotonos with hips and knees flexed and arms extended. He could say a few simple words, but speech was greatly inhibited by severe dystonic movements which involved vocalization. Relaxation was impossible. The child was very uncomfortable and sweated profusely. An encephalogram showed normal ventricles and subarachnoid markings. The spinal fluid was normal.

Operations. On May 17, 1949, an attempt was made to remove the right area 4, but because of inconclusive results on stimulation, the precentral gyrus was removed. The specimen measured 1 cm. in width and 1.5 cm. in depth.

Dystonia was abolished on the left side, and persisted on the right; opisthotonos persisted. The left extremities remained in plastic spasticity, with resistance chiefly to flexion.

On June 1, 1949, an attempt was made to excise the left area 4, but again this was thwarted by inconclusive results on stimulation. The precentral gyrus was excised in a cerebral sacrifice 4 cm. in diameter.

Thereafter no dystonia was noticed, opisthotonos did not recur and the child was comfortable. He had a spastic quadriplegia with the right arm flexed and the left extended, each resisting contrary passive motion. The legs were extended and crossed.

Course. These gains were maintained at examination 2 years later, except that the legs had become flexed. The child remained severely retarded mentally. He was well-nourished and comfortable. He had developed the ability to say a few words and could move the head and the extremities to a small extent.

Comment. This mentally retarded child’s dystonia involved the face, jaws and neck as well as the back and extremities. Removal of areas 4 and 6 (more extensively on the left) controlled the dystonia at the price of spastic quadriplegia and aphasia. The exchange was considered worthwhile by all involved in the care of the child.

Case 3. J. W., a 12-year-old white girl, had been born spontaneously after 7 months of gestation terminated by toxemia. The mother had experienced 7 idiopathic miscarriages.

The child was never able to hold up her head, sit, walk nor use her extremities. At the age of 4 she began having “torsion spasm.” Finally she was admitted because intermittent head retraction interfered with breathing, making suffocation appear imminent.

Examination. She was poorly nourished and developed. She lay in opisthotonos with the head retracted and turned to the right. Dystonic movements periodically increased the retraction, throwing into relief the hypertrophied sternomastoids over which coursed the distended jugular veins. As the movement was prolonged the face and chest became cyanotic. The child could be lifted from the floor by a hand under the occiput. Both arms usually were extended and the hands clenched. The left leg was extended and the right flexed. The left hip joint was dislocated and the ankle joint was flail-like. Dystonic and athetoid motion involved all extremities and was sharply increased by sensory stimuli and passive motion. The child apparently understood some words and spoke a very few with great effort. When relaxed she could move all extremities feebly and imperfectly.
An encephalogram revealed essentially normal ventricles and subarachnoid spaces.

Operations. On Nov. 12, 1951, the right cerebral peduncle was exposed and the anterior one-third of its lateral surface was incised to a depth of 5 mm. On Nov. 19, 1951, the left peduncle was incised similarly through the anterior one-half of its lateral surface.

Course. Postoperatively, the dystonia was abolished. The child could lie comfortably on her back; the neck was supple. The arms were maintained flexed, with the right wrist also flexed. Both legs were flexed, but could be extended within the limitation of her joint contractures. Both legs and arms resisted extension. The right arm also resisted flexion. The child spoke about as much as, but more easily than, before operation. She obviously was markedly deficient in mentality, as she had been before.

Comment. This child's dystonia involved all four extremities and the muscles of the back. Opisthotonos had progressed to a degree threatening suffocation by distortion of the airway. The dystonia and opisthotonos were abolished by interruption of the anterior one-half of the cerebral peduncles. While spastic quadriplegia resulted, the degree of original spasticity was reduced to the point of permitting return of some voluntary motion.

DISCUSSION

Data obtained from these 3 cases suggest that dystonic motion is mediated over both pyramidal and parapyramidal pathways (Fig. 1). It may be abolished at the cortical level by removal of areas 4 and/or 6, above the basal ganglia, by interruption of these pathways in the ventral halves of the cerebral peduncles or in the spinal cord.

In Case 1, gross removal of area 6, sparing area 4 (and, no doubt, a significant amount of adjacent area 6), resulted in abolition of dystonic motion in the contralateral side. Sufficient reflex spasm (i.e., tonic muscular contraction resulting from the application of a stretch-stimulus to a muscle by extending a joint) persisted to be disabling. This was abolished by the subsequent removal of area 4. Dystonia was abolished in the appropriate extremities in Case 2 by gross removal of the precentral gyrus alone.

Similar results followed interruption of these separate pathways in the spinal cord. Unilateral anterior column section abolished the dystonic movement in the leg, but the reflex spasm and some athetosis persisted. These were reduced markedly after pyramidal cordotomy. Unfortunately, the beneficial effects of cord sections are not permanent. Perhaps the impulses subserving the abnormal motion become re-routed over pyramidal and extrapyramidal pathways that escape section in the limited incisions that are possible in human subjects.

Bilateral partial cerebral pedunculotomy is the operation of choice of the three procedures. The results are more favorable and the side-effects are less disabling than are those following cortical operations. The favorable effects include more body area than do those of cord section.
Fig. 1. Diagrammatic representation of the course of the pyramidal and parapyramidal tracts, exclusive of the brain-stem connection.


These experiences are marked by too many variables to assist much in a better understanding of the mechanism of dystonia. The area 4 and 6 decortications were too gross to permit assay of the effects of precise lesions. The peduncle incisions are relative unknowns concerning specific tracts severed and the amounts included. As Meyers has noted, the concepts of anatomists vary widely as to the area and extent in the cross section of the peduncles of the several traversing fiber tracts. It is now appreciated that cross sectional area-maps of the spinal cord fiber tracts are only gross approximations, representing maximal concentrations, with great intermixtures. There is little reason to doubt that a similar state exists in the cerebral peduncles.

It is interesting to speculate upon the possible effects of making sections above and below the points of maximal intake of fibers connecting with the feed-back circuits of the reticular formation, the cerebellum and the basal ganglia. The present experiments show little difference in the motor effects of lesions made above and below the level of the basal ganglia. The results of interruption of fibers between the feed-back mechanisms and the motor cortex still remain to be demonstrated.
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ADDENDUM

Since submission of this manuscript, bilateral cerebral pedunculotomy has been performed on 2 additional children (L.S. and I.S.) for severe dystonia. The early results in these cases resemble those of J.W. (Case 3). The gains reported in the latter case have persisted to date, approximately 22 months.